Product Data Sheet: Frataxin

Cat. No:	MAB-10876
Conjugate:	Unconjugated
Size:	100 ug
Clone:	STR23
Concentration:	1mg/ml
Host:	Ms
Isotype:	IgG1,k
Immunogen:	Human frataxin (full length mature form, residues 56-210)
Reactivity:	Hu
Applications:	Western blotting , Immunocytochemistry, Flow Cytometry , IHC(P)
Purification:	Purified
Background:	Frataxin is a mitochondrial protein that is approximately 17 kDa in size in its monomeric form. The gene encoding frataxin in humans lies on chromosome 9. The codon GAA is repeated 100-1700 times in both copies of this gene in the majority of patients having Friedreich's Ataxia (FRDA). FRDA is an autosomal recessive disorder causing neurodegeneration and cardiomyopathy. FRDA patients show low expression of frataxin. Frataxin has been implicated in mitochondrial iron homeostasis although the mechanism by which this protein is involved in iron metabolism is still not clear. Deletion of the frataxin gene in yeast results in iron accumulation in the mitochondria and loss of respiration. Increased mitochondrial iron levels have also been observed in FRDA patients. Recombinant human frataxin, both in its monomeric form and in a polymeric form, has been shown to bind iron in vitro.
Form:	Liquid

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